

wherein:  $X^+$  is  $N^+(R_1, R_2, R_3)$ , wherein

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$R_1, R_2, R_3$ , being the same or different, are selected in the group consisting of hydrogen, a  $C_1$ - $C_9$  straight or branched alkyl group,  $-CH=NH(NH_2)$ ,  $-NH_2$ , and  $-OH$ ; or one or more  $R_1, R_2$  and  $R_3$ , together with the nitrogen atom which they are linked to, form a saturated or unsaturated, monocyclic or bicyclic heterocyclic system; with the proviso that at least one of the  $R_1, R_2$  and  $R_3$  is different from hydrogen;

*Sub C1*

$Z$  is selected from

*Sub C1*

- $-OR_4$ ,
- $-OCOOR_4$ ,
- $-OCONHR_4$ ,
- $-OCSNHR_4$ ,
- $-OCSOR_4$ ,
- $-NHR_4$ ,
- $-NHCOR_4$ ,
- $-NHCSR_4$ ,
- $-NHCOOR_4$ ,
- $-NHCSOR_4$ ,
- $-NHCONHR_4$ ,
- $-NHCSNHR_4$ ,
- $-NHSOR_4$ ,
- $-NHSONHR_4$ ,
- $-NHSO_2R_4$ ,

-NHSO<sub>2</sub>NHR<sub>4</sub>, and

-SR<sub>4</sub>,

wherein -R<sub>4</sub> is a C<sub>1</sub>-C<sub>20</sub> saturated or unsaturated, straight or branched alkyl group, optionally substituted with an A<sub>1</sub> group, wherein A<sub>1</sub> is selected from the group consisting of a halogen atom, or an aryl, heteroaryl, aryloxy or heteroaryloxy group, said aryl, heteroaryl, aryloxy or heteroaryloxy groups being optionally substituted with one or more C<sub>1</sub>-C<sub>20</sub> saturated or unsaturated, straight or branched alkyl or alkoxy group and/or halogen atom;

Y<sup>-</sup> is selected from the group consisting of -COO<sup>-</sup>, PO<sub>3</sub>H<sup>-</sup>, -OPO<sub>3</sub>H<sup>-</sup>, tetrazolate-5-yl;

with the proviso that when Z is -NHCOR<sub>4</sub>, Y is -COO<sup>-</sup>, then R<sub>4</sub> is C<sub>20</sub> alkyl;

with the proviso that when Z is -NHSO<sub>2</sub>R<sub>4</sub>, Y<sup>-</sup> is -COO<sup>-</sup>, then R<sub>4</sub> is not tolyl;

with the proviso that when Z is -NHR<sub>4</sub>, X<sup>+</sup> is trimethylammonium and Y<sup>-</sup> is -COO<sup>-</sup>, then R<sub>4</sub> is not C<sub>1</sub>-C<sub>6</sub> alkyl,

their (R,S) racemic mixtures, their single R or S enantiomers, or their pharmaceutically acceptable salts .

2 29. (New) A compounds according to claim 28, wherein R<sub>1</sub>, R<sub>2</sub> and R<sub>3</sub> are methyl.

30. (New) A compounds according to claim 28, wherein the heterocyclic system formed by R<sub>1</sub>, R<sub>2</sub> and R<sub>3</sub> together with nitrogen is selected from the group consisting of morpholinium, quinuclidinium, pyridinium, quinolinium and pyrrolidinium.

31. (New) A compound according to claim 28, wherein R<sub>1</sub> and R<sub>2</sub> are H, R<sub>3</sub> is selected from the group consisting of -CH=NH(NH<sub>2</sub>), -NH<sub>2</sub> and -OH.

32. (New) A compound according to claim 28, wherein Z is selected from the group consisting of ureido (-NHCONHR<sub>4</sub>) or carbamate (-OCONHR<sub>4</sub>), and R<sub>4</sub> is a C<sub>7</sub>-C<sub>20</sub> saturated or unsaturated, straight or branched alkyl group.

33. (New) A compound according to claim 32, wherein R<sub>4</sub> is a C<sub>9</sub>-C<sub>18</sub> saturated or unsaturated, straight or branched alkyl group.

34. (New) A compound selected from the group consisting of

R,S-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;

R,S-4-quinuclidinium-3-(tetradecyloxycarbonyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(nonylcarbamoyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(nonyloxycarbonyl)-oxybutyric acid chloride;

R,S-4-trimethylphosphonium-3-(nonylcarbamoyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(octyloxycarbonyl)-aminobutyrate;

R,S-4-trimethylammonium-3-(nonyloxycarbonyl)-aminobutyrate;

R,S-4-trimethylammonium-3-octyloxybutyrate;  
R,S-4-trimethylammonium-3-tetradecyloxybutyrate;  
R,S-1-guanidinium-2-tetradecyloxy-3-(tetrazolate-5-yl)-propane;  
R,S-4-trimethylammonium-2-tetradecyloxy-3-(tetrazolate-5-yl)-propane;  
R,S-3-quinuclidium-2-(tetradecyloxycarbonyl)-oxy-1-propanephosphonate  
monobasic;  
R,S-3-trimethylammonium-2-(nonylaminocarbonyl)-oxy-1-  
propanephosphonate monobasic ;  
R,S-3-pyridinium-2-(nonylaminocarbonyl)-oxy-1-propanephosphonic acid  
chloride;  
R-4-trimethylammonium-3-(tetradecylcarbamoyl)-aminobutyrate;  
R-4-trimethylammonium-3-(undecylcarbamoyl)-aminobutyrate;  
R-4-trimethylammonium-3-(heptylcarbamoyl)-aminobutyrate;  
R,S-4-trimethylammonium-3-(nonylthiocarbamoyl)-aminobutyrate;  
R-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;  
S-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;  
S-4-trimethylammonium-3-(tetradecylcarbamoyl)-aminobutyrate;  
R,S-4-trimethylammonium-3-tetradecylaminobutyrate;  
R,S-4-trimethylammonium-3-octylaminobutyrate;  
R,S-4-trimethylammonium-3-(decansulfonyl)aminobutyrate;  
R,S-4-trimethylammonium-3-(nonylsulfamoyl)aminobutyrate;  
S-4-trimethylammonium-3-(dodecansulfonyl)aminobutyrate;  
R-4-trimethylammonium-3-(dodecansulfonyl)aminobutyrate;

S-4-trimethylammonium-3-(undecylsulfamoyl)aminobutyrate;  
R-4-trimethylammonium-3-(undecylsulfamoyl)aminobutyrate;  
R-4-trimethylammonium-3-(dodecylcarbamoyl)aminobutyrate;  
R-4-trimethylammonium-3-(10-phenoxydecylcarbamoyl)aminobutyrate; and  
R-4-trimethylammonium-3-(*trans*- $\beta$ -styrenesulfonyl)aminobutyrate.

8 35. (New) A process for the preparation of a compound of claim 28,  
wherein Z is carbonate (-OCOOR<sub>4</sub>), carbamate (-NHCOOR<sub>4</sub>), thiocarbamate (-  
OCSNHR<sub>4</sub>) or thiocarbonate (-OCSOR<sub>4</sub>), said process comprising reacting X<sup>+</sup>-  
CH<sub>2</sub>-CH(OH)-CH<sub>2</sub>-Y<sup>-</sup>, of the desired structure, optionally protected on the acid  
Y<sup>-</sup> group, respectively with an alkyl chloroformate, alkyl isocyanate, alkyl  
isothiocyanate or alkyl thiochloroformate, wherein the alkyl moiety is the  
desired R<sub>4</sub> alkyl group, to produce the desired compound.

9 36. (New) A process for a preparation of a compound of claim 28,  
wherein Z is amide (-NHCOR<sub>4</sub>), thioamide (-NHCSR<sub>4</sub>), carbamate (-NHCOOR<sub>4</sub>),  
thiocarbamate (-NHCSOR<sub>4</sub>), ureido (-NHCONHR<sub>4</sub>), thioureido (-NHCSNHR<sub>4</sub>),  
sulfinamide (-NHSOR<sub>4</sub>), sulfonamide (-NHSO<sub>2</sub>R<sub>4</sub>), sulfinamoylamino  
(-NHSONHR<sub>4</sub>), and sulfamide (-NHSO<sub>2</sub>NHR<sub>4</sub>), said process comprising reacting  
X<sup>+</sup>-CH<sub>2</sub>-CH(OH)-CH<sub>2</sub>-Y<sup>-</sup>, of the desired structure, optionally protected on the  
acid Y<sup>-</sup> group, respectively with an acyl chloride, thioacyl chloride, alkyl  
chloroformate, alkyl thiochloroformate, alkyl isocyanate, alkyl thioisocyanate,  
alkyl sulfinyl chlorides, alkyl sulfonyl chlorides, SOCl<sub>2</sub> and alkyl amines, alkyl

sulfamoyl chloride or  $\text{SOCl}_2$  and alkyl amine, wherein the alkyl moiety is the desired  $\text{R}_4$  alkyl group, to produce the desired compound.

10 37. (New) A process for the preparation of a compound of claim 28, wherein Z is  $-\text{OR}_4$  or  $-\text{SR}_4$ , said process comprising the steps of:

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- (a) reacting a carbonyl compound of formula  $\text{Hal-CH}_2\text{-CO-CH}_2\text{-COOR}'$ , wherein Hal is a halogen atom and  $\text{R}'$  is the residue of a suitable ester, with respectively alcohols and thiols  $\text{R}_4\text{OH}$  or  $\text{R}_4\text{SH}$ , to give the respective ketal or thioketal;
  - (b) transforming the respective ketal or thioketal into the respective ether or thioether;
  - (c) substituting the Hal atom with an azido group, and
  - (d) transforming the azido group into the  $\text{X}^+$  group to produce the desired compound.

11 38. (New) A process for the preparation of a compound of claim 28, wherein Z is  $-\text{NHR}_4$ , said process comprising reacting of  $\text{X}^+\text{-CH}_2\text{-CH(NH}_2\text{)-CH}_2\text{-Y}^-$  of the desired structure, optionally protected on the acid  $\text{Y}^-$  group, with alkane carbaldheydes, wherein the alkyl moiety is a one-term lower homologue of the desired  $\text{R}_4$ , and subsequent reduction, to produce the desired compound.

12 <sup>39</sup>. (New) A pharmaceutical composition comprising a therapeutically effective amount of a compound of claim 28<sup>1</sup>, in admixture with a pharmaceutically acceptable vehicle or and excipient.

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cmt 13 <sup>40</sup>. (New) The pharmaceutical composition according to claim 39<sup>12</sup>, wherein an active ingredient suitable for the treatment of diabetes is also present and is selected from the group consisting of sulfonylurea, L-carnitine, fibrate and other agonists of peroxisomal proliferator activated receptor (PPAR- $\alpha$ ), HMG-CoA reductase inhibitor,  $\beta$ -sitosterol inhibitor, cholesterol acyltransferase inhibitor, biguanides, cholestyramine, angiotensin II antagonist, melinamide, nicotinic acid, fibrinogen receptor antagonists, aspirin,  $\alpha$ -glucosidase inhibitors, insulin secretagogue, insulin and glucagon-like peptides and agonists of PPAR- $\gamma$ .

14 <sup>41</sup>. (New) A pharmaceutical composition according to claim 39<sup>12</sup>, also including an active ingredient suitable for the treatment of obesity selected from the group consisting of fenfluramine, dexfenfluramine, phentiramine, and a  $\beta$ -3-adrenergic receptor agonist.

15 <sup>42</sup>. (New) A pharmaceutical composition according to claim 39<sup>12</sup>, also including an active ingredient suitable for the treatment of high cholesterol levels and in modulating HDL plasma levels, which is selected from the group consisting of fibrates, and other PPAR- $\alpha$  agonists; inhibitors of cholesterol

biosynthesis, HMG-CoA reductase inhibitors, statins, inhibitors of cholesterol absorption, acyl CoA:cholesterol acyltransferase inhibitors, anion exchange resins, nicotinyl alcohol, nicotinic acid or a salt thereof, vitamin E, thyromimetics and L-carnitine.

16 43. (New) A method for treating a subject having hyperactive carnitine palmitoyl-transferase comprising administering to said subject an effective amount of a compound of claim 28. |

17 44. (New) A method for treating a subject having hyperglycaemia, diabetes, heart failure or ischemia comprising administering to said subject an effective amount of a compound of claim 28. |

18 45. (New) A method for treating a subject having obesity comprising administering to said subject an effective amount of a compound of claim 28. |

19 46. (New) A method for treating a subject having high triglyceridemia comprising administering to said subject an effective amount of a compound of claim 28. |

20 47. (New) A method for treating a subject having hypertension comprising administering to said subject an effective amount of a compound of claim 28. |



2) 48. (New) A method of modulating high cholesterol levels or MDL

plasma levels in a subject in need of same, said method comprising

administering to said subject an effective amount of a compound of claim 28.

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